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CASE FOR DIAGNOSIS

Recurrent Reticulated Reddish-Orange Rash on the Neck and Inframammary Regions

Erupción reticulada eritematoanaranjada recurrente en cuello y región inframamaria

Clinical History

A 26-year-old white woman with no personal or family history of interest was referred to our department for a highly pruritic rash on the back of the neck, upper part of the back, and inframammary region (Figure 1, A and C). The rash had developed 3 months earlier and was resistant to treatment with topical and oral corticosteroids and oral antihistamines. The patient stated that she had had a similar rash some months before this episode and that it had resolved spontaneously in a few weeks.

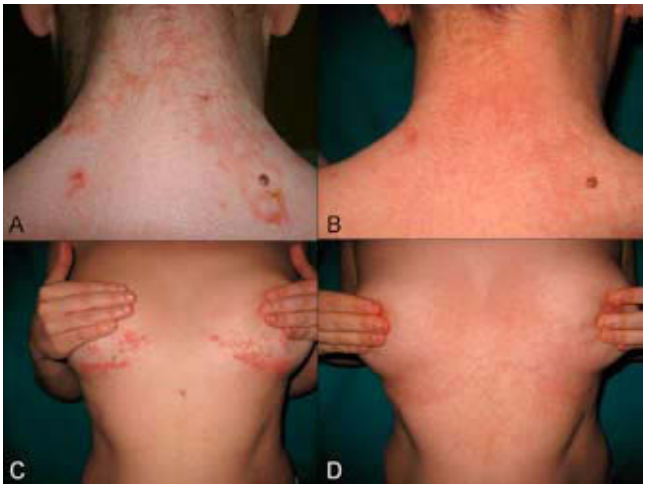

Figure 1

Physical Examination

On physical examination there was a rash of confluent erythematous papules of urticarial appearance in the areas described. There were signs of scratching. Some of the papules had become vesicular and subsequently developed erosions that left scabs (Figure 1, A and C).

Histopathology

Histopathology of a punch biopsy of one of the lesions revealed a perivascular mixed inflammatory infiltrate in the reticular and papillary dermis, formed mainly of lymphocytes and polymorphonuclear cells. In addition, there was exudation of polymorphonuclear cells into the epidermis and multiple necrotic keratinocytes, some grouped in foci that, in combination with the spongiotic changes, gave rise to true intraepidermal vesicles (Figure 2).

Additional Tests

The results of a complete blood count, biochemistry, and antinuclear antigens were normal.

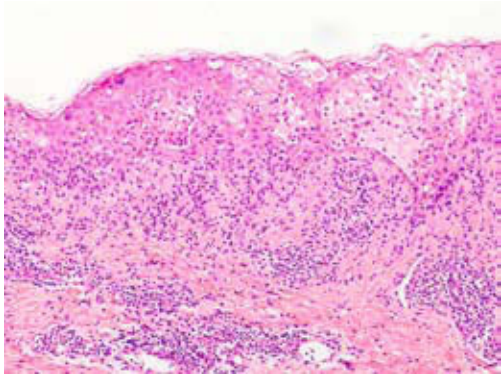

Figure 2 Hematoxylin-eosin, original magnification ×40.

What Is Your Diagnosis?

Diagnosis

Prurigo pigmentosa

Clinical Course

The patient returned 3 weeks later and had no active lesions, with only a residual reticular pigmentation in the areas previously affected (Figure 1, B and D). It was decided to continue observation and to prescribe treatment if there was a further recurrence. No new lesions had appeared after 7 months.

Discussion

Prurigo pigmentosa is an uncommon inflammatory skin disease of unknown etiology that mainly affects young adult women, particularly Japanese women,[1] although it has also been reported in men. To date it has not been reported in prepubertal children or in the elderly.[2] It consists of a recurrent pruritic rash of erythematous papules, vesicles, and scabs that resolves leaving a reticular or mottled pigmentation. It is typically distributed symmetrically on the neck, upper part of the back, and inframammary and clavicular regions, although it sometimes also affects the abdomen, lumbosacral region, and face. The mucous membranes are spared.[3]

The rash tends to recur in the same areas that have previously been affected.[4] The etiology and pathogenesis are unknown, although a number of possible mechanisms have been suggested, including friction, contact allergy, exposure to sunlight, various endocrine disturbances (eg, diabetes mellitus) and metabolic disorders (eg, ketosis),[1] pregnancy, and anorexia nervosa,[3] though no consistent relationship has been demonstrated.[1]

Diagnosis requires correlation of the clinical and pathology findings.[5] Histopathology in the early stages of the disease reveals a superficial perivascular neutrophilic infiltrate, nuclear fragmentation, edema of the papillary dermis, and a degree of neutrophilic spongiosis in the epidermis. As the disease develops, frank spongiotic vesicles appear, associated with neutrophilic microabscesses, keratinocyte necrosis, a lymphocytic infiltrate in a lichenoid distribution, dyskeratotic cells, and a number of eosinophils in the dermis and epidermis; there may also be intraepidermal and subepidermal blisters.[2]

In the initial inflammatory phase, the differential diagnosis includes dermatitis herpetiformis, linear immunoglobulin A dermatosis, and acute cutaneous lupus erythematosus. In the residual pigmentary phase, confluent and reticulated papillomatosis of Gougerot-Carteaud, congenital dyskeratosis, Dowling-Degos disease, lichen planus pigmentosus, pigmented contact dermatitis, erythema dyschromicum perstans, and macular amyloidosis must be considered.[2]

The treatments that have been found to achieve best results are minocycline (100-200 mg/d) and dapsone (25-100 mg/d), administered for several weeks[1-6]; with these treatments, the pruritus and rash disappear, but the residual reticular pigmentation is not prevented. Some authors have suggested that minocycline is more effective and safer than dapsone.[6] The mechanism of action of these drugs appears to involve inhibition of neutrophil migration or function. Other drugs that have been used to treat this skin disease include isotretinoin,[5] doxycycline,[5,6] and sulfamethoxazole. The macrolides have also been proposed as a therapeutic option.[4]

Conflict of Interest

The authors declare that they have no conflict of interest.

Acknowledgments

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